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Case report

Gallbladder Agenesis

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ABSTRACT

Gallbladder agenesis is a very rare anomaly which can cause many diagnostic and therapeutic dilemmas. About 20% of patients present with right upper quadrant pain, nausea and fatty food intolerance. This condition is frequently unrecognized and leads to unnecessary and potentially dangerous surgical procedures. We present a case report of a 46-year old woman who had clinical presentation of symptomatic cholelithiasis. The diagnosis of cholelithiasis was verified ultrasonographically. We decided to perform laparoscopic cholecystectomy but during surgery gallbladder was not found. Laparoscopic procedure was therefore stopped and 7 days postoperatively MR Cholangiopancreatography was performed – it was clearly that patient had gallbladder agenesis.

Keywords: *gallbladder, laparoscopy, gallstones, pathology, agenesis*

Introduction

Gallbladder agenesis (GA) is one of the several gallbladder congenital abnormalities[1]: absence of the gallbladder, duplication of the gallbladder, deformation of the gallbladder, left-sided gallbladder, intrahepatic gallbladder and mobile gallbladder. Gallbladder agenesis was first described in 1702 by Bergman[2].

It is a very rare malformation (0.01%–0.065% of the population) and develops from the failure of normal development of cystic bud in utero. Women-men ratio is 3:1 and can be associated with other malformations like cardiovascular, musculoskeletal and gastrointestinal abnormalities[3].

About 20% of patients with GA become symptomatic (right upper abdominal pain, nausea, vomiting or fatty food intolerance). Gold standard for diagnosis of gallstone disease is ultrasound (US). GA can be misinterpreted as a shrunken, scarred and atrophic gallbladder. Some of these patients are scheduled for laparoscopic cholecystectomy – which is unnecessary and potentially dangerous surgical procedure.

Case report

A 46-year old woman presented to our Department with 1 year history of intermittent right upper abdominal pain. Abdominal ultrasound showed signs of chronic cholecystitis with intraluminal lithiasis (Figure 1). During clinical examination abdomen was soft, without tenderness, and Murphy sign was negative. Her vital signs were normal. In the laboratory findings she had hypercholesterolemia with normal other biochemical findings, including bilirubin and liver enzymes. She was scheduled for laparoscopic cholecystectomy. During surgical procedure we prepared d.choledochus which was in normal range, but gallbladder was not showed (Figure 2). Surgeon stopped procedure because of high possibility of injuring bile structures. To evaluate the intraoperative findings magnetic resonance cholangiopancreatography (MRCP) was performed (Figure 3). MRCP confirmed the absence of the gallbladder and cystic duct (Figures 4,5). The normal diameter of common bile duct and intrahepatic bile ducts was found.

Discussion

Gallbladder is formed in the fourth week of intrauterine life and arises from the caudal bud of the hepatic diverticulum along with the cystic duct and the ventral pancreas (4). What is the reason of the gallbladder agenesis is unknown, but there are two hypotheses. First of them postulates that the superior division of the caudal

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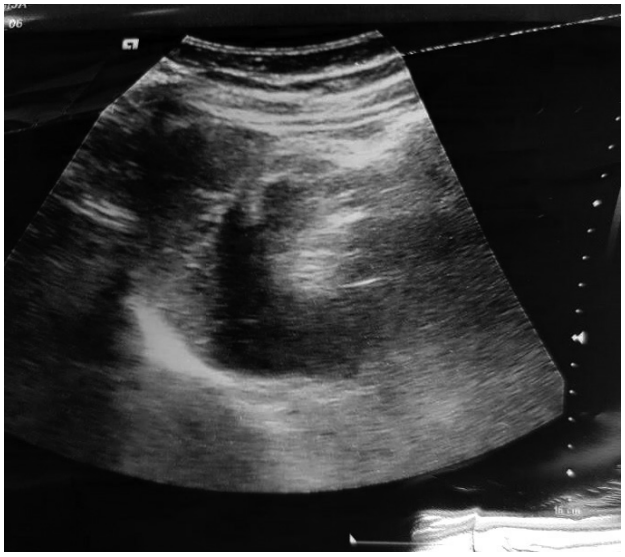

Fig 1. US showed chronic cholecystitis

Fig 3. MRI T2W - absence of the gallbladder.

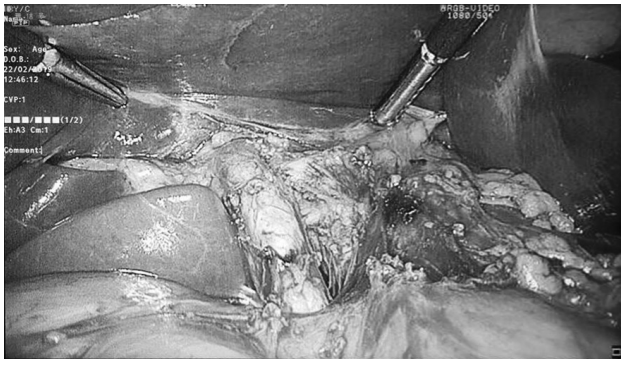

Fig 2. Intraoperative findings – common bile duct, without gallbladder.

Fig 4. 3D MRCP- confirmed the diagnosis of the gallbladder agenesis.

Fig 5. Reconstruction MRCP- volume rendering technique - gallbladder agenesis.

bud of the hepatic diverticulum fails to develop into the cystic duct and gallbladder, possibly secondary to an aberration in the development of the vessels surrounding the caudal bud[5,6,7]. The second theory tells that failure of the recanalisation of the gallbladder and cystic duct after their formation causes gallbladder agenesis[8]. Patients with GA are classified by Bennion et al. into 3 groups: the first group are patients with multiple fetal anomalies, including GA, and all of them died secondarily to their other congenital defects. Second group is the asymptomatic group of patients in whom GA was diagnosed post mortem. A third group are the patients with gastrointestinal symptoms who resulted with the intraoperative diagnosis[9].

About 50–70% of patients with GA is asymptomatic. Symptomatic patients usually present with chronic right upper quadrant pain, dyspepsia, jaundice and fatty food intolerance. Those symptoms are similar to symptoms experienced in acute cholecystitis or biliary colic. Explanation for these symptoms in patients with GA is that patients can have sphincter of Oddi dysfunction which causes biliary stasis and following biliary stasis jaundice, elevated liver function tests and common bile duct stones[10,11].

Preoperative diagnosis of the GA is very challenging, and many of these patients undergo unnecessary surgical procedure- laparoscopic cholecystectomy[12,13]. Dissection of the falciform ligament, common bile duct and other anatomical structures of the hepatobiliary tree to locate gallbladder is very dangerous and can cause many complications. Intraoperative cholangiography and intraoperative US can be helpful to resolve this intraoperative dilemma[14].

Non visualisation of the gallbladder preoperatively can be interpreted by obstruction of the cystic duct or with a contracted gallbladder.

In our case, MRCP was done postoperatively, and we confirmed the diagnosis of the GA[15,16]. Some authors recommend HIDA (iminodiacetic acid) hepatobiliary scan or ERCP[17]. In our opinion MRCP is a better diagnostic tool than endoscopic retrograde cholangiopancreatography (ERCP) because it is not an invasive technique and confirms very clearly diagnosis of the GA.

Conclusion

Gallbladder agenesis is a very rare anatomical variation of the hepatobiliary tree, but in some cases can cause many diagnostic and intraoperative dilemmas. In our opinion, non-visualisation of the gallbladder during laparoscopic cholecystectomy should not lead to the open surgical procedure especially when no other known pathology in the biliary tree is present. In such cases we recommend postoperative diagnostic tools for confirming the diagnosis of the GA.

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AGENEZA ŽUČNJAKA

SAŽETAK

Ageneza žučnjaka je vrlo rijetka anomalija ali može prouzročiti brojne dijagnostičke i terapijske dileme. Oko 20% pacijenata sa agenezom žučnjaka ima simptome boli u gornjem desnom abdominalnom kvadrantu, mučnine i intoleranciju masne hrane. Takvi simptomi vrlo često mogu voditi do nepotrebnih i potencijalno opasnih dijagnostičko-terapijskih postupaka. Želimo prezentirati naš slučaj, 46-godišnje žene sa kliničkom prezentacijom simptomatske kolelitijaze koja je potvrđena ultrazvučnim pregledom. Tijekom laparoskopske kolecistektomije nismo mogli naći žučnjak te smo odlučili prekinuti operativni zahvat, da bi konačnu dijagnozu utvrdili postoperativno, uz pomoć MRCP-a.